Organ of Chievitz: Revisited
Mamata S Kamat, Rudrayya Puranik, Shrinivas Vanaki, Varsha VK

Abstract
Organ of Chievitz is a normal, epithelial structure of the oral cavity located in the most postero medial aspect of medial surface of ramus of mandible. It consists of benign epithelial islands or nests. In the current article, the location, embryology, histological appearance and diagnostic workup of JOC are revisited which is of special interest to the surgical pathologist for the oral cancer diagnosis, as they are in the potential of being misinterpreted as carcinoma or perineural invasion particularly on frozen sections. The purpose of the present article is to update the clinicians about the recent concepts and diagnostic pitfalls so that unnecessary investigations and overtreatment is prevented for the betterment of patients.

Key words: Carcinoma; Cytokeratin; Juxtaoral organ of Chievitz; Perineural

Introduction
Juxta oral Organ of Chievitz (JOC) first described by Chievitz in 1885 is a normal permanent epithelial structure intimately associated with nerves found in the soft tissue overlying the angle of mandible in the buccotemporal space. It is located typically between the temporalis muscle and the buccinator, medial to the mandibular internal oblique ridge close to the site of injection for inferior alveolar nerve block. Anterior extremity is behind and below the area where stenson’s duct penetrates the buccinator muscle. Anatomic location of JOC is depicted in figure 1. It is microscopic and fusiform in structure, measuring between 7-15 mm in length, 1-2 mm in diameter and not normally palpable. They may present as benign nodular hyperplasia. Histologically, they show epithelial islands made of benign squamous cells with cuboidal basal cells at the periphery. This paper throws light on the salient features of JOC with recent views on origin and emphases on differential diagnosis, so that overtreatment is avoided.

History and Theories for development of JOC
a) Chievitz (1885) thought JOC as remnants of salivary glands which disappear before birth. Strong activity of hydrolase, alkaline phosphatase and carbonic anhydrase enzymes by the epithelial component suggested it to be an abortive salivary gland.

b) Ramsay (1935) supported the theory of oral epithelial inclusions trapped by the fusion of maxillary and mandibular processes and no evidence of either endocrine or exocrine functions, presumed to disappear with maturity.

c) Zenker et al (1953 -1969) opined that these organs persisted in the adult and appeared to be chemically active supporting a neuroendocrine receptor organ theory and named them as Juxta oral organ.

d) Lutman (1974) reported presence of benign epithelial nests associated with sensory nerve fibres and supported Ramsay theory.

e) Recently Ischen and Fechner (1979) suggested that these organs are not associated with oral carcinoma.

f) Some authors consider JOC to be a hamartomatous proliferation.

g) Recently reports suggest that the cells of JOC are of neural origin and does not represent a neuroreceptor.

Functions of JOC
Some authors suggest that these organs are not of functional importance and are vestigial. Others opined that these have a secretory activity (secretion of unknown secretory product) and neuroreceptor function involved in perception during suckling, swallowing, mastication and speech.

Histology
Microscopically it shows multiple (two to ten) well circumscribed, multilobulated mass of
discrete cell nests resembling squamous epithelium. There is no evidence of keratinisation. The cell nests are enclosed sharply by basement membrane in a fibrous stroma. The cells contain pink to clear cytoplasm with variably sized nuclei ranging from vesicular to hyperchromatic. Palisading of nuclei along the basement membrane is also seen sometimes. Some cell nests show lumens filled with mucin negative secretions and exhibit focal calcifications. There is no evidence of keratinisation. The cell nests are enclosed sharply by basement membrane in a fibrous stroma. The cells contain pink to clear cytoplasm with variably sized nuclei ranging from vesicular to hyperchromatic. Palisading of nuclei along the basement membrane is also seen sometimes. Some cell nests show lumens filled with mucin negative secretions and exhibit focal calcifications. The stroma that surrounds epithelial islands is typically loose and fibrous (stratum fibrosum internum) which in turn is surrounded by a dense connective tissue layer (stratum fibrosum externum). These islands are usually associated with sensory nerve.


Stains for demonstration of JOC: Routine Haematoxylin and Eosin, Periodic Acid Schiff for basement membrane, mucicarmine for mucin and von Kossa for calcifications.

Immunohistochemical Profile: All the epithelial cells of JOC show diffuse and strong positivity for CKs (AE1/AE3, 34βE12 and MNF 116). Exclusive expression of CK14 is seen among the central squamous cells. More intense expression of CK10 and CK19 is seen in central squamous cells than peripheral basal cells. The cells showed negative expression for CK13, CK18, filagrin, involucrin and epithelial membrane antigen. These features suggest that the epithelial cell nests of JOC are of non-keratinised stratified squamous cells. Some studies have suggested that the cells are positive for Vimentin, S-100 protein, Neuron-specific enolase, neural cell adhesion molecule and high and low affinity nerve growth factor receptors. This suggests that the cells of JOC are of neural origin. Lack of positive reaction for chromogranin, synaptophysin, neurofilament and protein gene product 9.5 around the Chievitz cell nests indicate that they do not represent neuroreceptor.

Differential Diagnosis
JOC needs to be differentiated from Odontogenic tumors, Perineural invasion of squamous cell carcinoma, adenocarcinoma and mucoepidermoid carcinoma. • JOC versus Perineural carcinomatous invasion:
  a) the epithelial islands of JOC are surrounded by basement membrane while malignant cells are not,
  b) two distinct layers connective tissue are evident in JOC and not in carcinoma involving nerve,
  c) the epithelial cells of JOC do not abut directly on the nerves, whereas malignant cells invade or touch the perineurium in perineural invasion,
  d) significant pleomorphism and mitosis are not seen in JOC in contrast to carcinomas,
  e) desmoplastic inflammatory stromal reaction is often associated with invasive carcinoma but not with JOC,
  f) according to Mandle, JOC is immunoreactive with Anti CK 19
Antibodies, whereas neoplastic cells are not.

- JOC versus adenocarcinoma, mucoepidermoid carcinoma: although glandular foci filled with colloid are sometimes seen in JOC, the lack of mucin stain, helps to differentiate it from adenocarcinoma and mucoepidermoid carcinoma.

- JOC versus odontogenic tumors: location of JOC (behind the tooth-bearing area, deep in the medial pterygoid muscle, at the level of the pterygomandibular raphe) aid in differentiating it from odontogenic tumors (odontogenic epithelial cell rests can be found around tooth-bearing areas).

Conclusion
These features should enlighten the surgical pathologists regarding the development, location and histological appearance of JOC to avoid serious overtreatment and unnecessary radical surgery resulting in high morbidity of the patient. Further it also necessitates the need to report these structures to widen the knowledge regarding the true nature of this organ.

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